

University of Groningen

## Perceived competence in children and adolescents with haemophilia

Hegeman, A. K.; van Genderen, F. R.; Meijer, S.; van den Briel, M. M.; Tamminga, R. Y. J.; van Weert, E.

*Published in:*  
Haemophilia

*DOI:*  
[10.1111/j.1365-2516.2010.02357.x](https://doi.org/10.1111/j.1365-2516.2010.02357.x)

**IMPORTANT NOTE:** You are advised to consult the publisher's version (publisher's PDF) if you wish to cite from it. Please check the document version below.

*Document Version*  
Publisher's PDF, also known as Version of record

*Publication date:*  
2011

[Link to publication in University of Groningen/UMCG research database](#)

### *Citation for published version (APA):*

Hegeman, A. K., van Genderen, F. R., Meijer, S., van den Briel, M. M., Tamminga, R. Y. J., & van Weert, E. (2011). Perceived competence in children and adolescents with haemophilia: an explorative study. *Haemophilia*, 17(1), 81-89. <https://doi.org/10.1111/j.1365-2516.2010.02357.x>

### **Copyright**

Other than for strictly personal use, it is not permitted to download or to forward/distribute the text or part of it without the consent of the author(s) and/or copyright holder(s), unless the work is under an open content license (like Creative Commons).

The publication may also be distributed here under the terms of Article 25fa of the Dutch Copyright Act, indicated by the "Taverne" license. More information can be found on the University of Groningen website: <https://www.rug.nl/library/open-access/self-archiving-pure/taverne-amendment>.

### **Take-down policy**

If you believe that this document breaches copyright please contact us providing details, and we will remove access to the work immediately and investigate your claim.

*Downloaded from the University of Groningen/UMCG research database (Pure): <http://www.rug.nl/research/portal>. For technical reasons the number of authors shown on this cover page is limited to 10 maximum.*



## ORIGINAL ARTICLE *Paediatrics*

# Perceived competence in children and adolescents with haemophilia: an explorative study

A. K. HEGEMAN,\* F. R. VAN GENDEREN,§<sup>1</sup> S. MEIJER,\* M. M. VAN DEN BRIEL,\* R. Y. J. TAMMINGA† and E. VAN WEERT\*‡

\*Center for Rehabilitation; †Beatrix Children's Hospital; ‡SHARE-GSMS, Graduate School of Medical Sciences, University Medical Center Groningen, University of Groningen, Groningen, The Netherlands; and §Department of Pediatric Physical Therapy and Clinical Exercise Physiology, University Children's Hospital, UMC Utrecht and Van Creveldkliniek, UMC Utrecht, The Netherlands

**Summary.** With the introduction of prophylaxis, restricting children with haemophilia to participate in physical activities was no longer necessary. Subsequently, many studies report on improved physical functioning in children and adolescents with haemophilia. However, little is known about psychological aspects such as perceived competence and impact of disease. Therefore, the aims of this study were to explore: (i) perceived competence, (ii) perceived impact of illness, and (iii) analyse associations between perceived competence and demographic factors, disease-related factors and joint status in young haemophiliacs in the Netherlands. Fifty-four children (age 8–12 years) and 72 adolescents (12–18 years) with haemophilia participated in this cross-sectional, multi-centre, explorative study. Measurements included perceived competence (Self-Perception Profile for Children/Adolescents; range 6–24/5–20), impact of disease (Revised Perception Illness Experience; range 1–5), demographic factors, disease-related factors, joint status and functional status. Mean (SD) scores for perceived competence in the children ranged from 17.3 ( $\pm 4.0$ ) to 19.6 ( $\pm 4.0$ ), and for adoles-

cents from 13.3 ( $\pm 2.4$ ) to 15.7 ( $\pm 2.8$ ) points. In general, scores were comparable with those of healthy peers, but children with haemophilia had a lower global self-worth score and competence in close friendship was lower for adolescents when compared with those of healthy peers. Mean (SD) scores for impact of disease ranged from 1.2 ( $\pm 0.4$ ) to 2.3 ( $\pm 0.8$ ) in children and from 1.3 ( $\pm 0.4$ ) to 2.0 ( $\pm 0.8$ ) in adolescents. Severe haemophilia, prophylactic medication, high impact of disease and a shorter walking distance showed a weak to moderate association with perceived competence. Children and adolescents with haemophilia in general have a perceived competence that is nearly comparable with that of healthy peers, with the exception of a lower global self-worth in children and a lower competence for close friendship in adolescents. Haemophiliacs seem to perceive their disease as having relatively low impact on their life. Severe disease, prophylactic treatment and low functional status seemed to be associated with lower perceived competence.

**Keywords:** functional status, haemophilia, impact of disease, perceived competence

## Introduction

Haemophilia is a life-threatening and lifelong condition caused by a deficiency of coagulation factors VIII or IX. Resulting from this deficiency are the most important clinical characteristics of haemophilia, intramuscular

and intra-articular bleeds. Frequent bleeding can lead to joint degeneration, pain, functional limitations and subsequent disablement. Before the introduction of factor replacement therapy in the 1960s, haemophiliacs, especially children, were restricted in their physical activities in order to prevent joint bleeds. With the introduction of prophylaxis and home treatment, the restrictions were relaxed and boys with haemophilia were able to lead a more or less normal life, both physically and socially [1]. However, recent studies on outcome are not consistent. Some reported a good joint health and no limitations of activities in children with haemophilia [2], whereas others found minor impairments in joint health, good functional ability and

Correspondence: Anneke Hegeman, Center for Rehabilitation, University Medical Center Groningen, University of Groningen, PO Box 30001, 9700 RB Groningen, The Netherlands.  
Tel.: +31 50 361 0297; fax: +31 50 361 1707;  
e-mail: a.k.hegeman@rev.umcg.nl

<sup>1</sup>Present address: Baxter B.V., Utrecht, The Netherlands.

Accepted after revision 2 June 2010

muscle strength [3]. Additional studies reported a decreased physical functioning, i.e. lower muscle force [4,5] and (an)aerobic capacity [3,4,6]. Moreover, children with haemophilia reported a high impact of the disease on their lives, despite a normal physical function [7]. It was therefore recommended to include in future research both physical and psychosocial aspects when assessing the well-being of patients with haemophilia [7].

One of the psychological aspects may be a patient's perceived competence. Perceived competence refers to two types of cognitions [8]. The first is the global self-worth, which reflects the overall value one places on the self as a person. The second refers to domain-specific evaluations of one's competence or adequacy, such as competence in the cognitive, social and physical domains [9]. Perceived competence is influenced by native intrinsic competence needs and stimulated by a responsive environment [10]. In general, those with positive self-worth are typically better able to face the challenges of daily life.

The importance of perceived competence has been recognized in the field of paediatric chronic diseases. Several studies reported on associations between chronic disease such as congenital heart disease [11], musculoskeletal pain [12], epilepsy [13], asthma [13] and cerebral palsy [14] and psychosocial problems and maladjustment. Such studies revealed that the type of condition may have specific identifiable effects on psychosocial problems such as perceived competence beyond the general effect of living with chronic conditions.

A chronic condition such as haemophilia may also negatively influence an individual's perceived competence. For instance, boys with haemophilia might – in response to pain or bleeding – withdraw from regular activities and social contacts, and may therefore have fewer opportunities to succeed, which may hypothetically affect their self-perception. In addition, protective measures issued by a parent or professional may also limit participation in several activities, thereby decreasing the global self-worth or the domain-specific competence of young haemophiliacs, even if they are not suffering from acute bleedings. Moreover, the presence of a disease such as haemophilia increases the risk of setting children apart from peers both physically and socially. To our knowledge, only one study examined perceived competence in children with haemophilia and revealed a lower global self-worth and lower scholastic and athletic competence [15]. However, that study was based on a small sample size, included an American population and used the same instrument for children and adolescents while it is recommended to use age-specific instruments [16]. To our knowledge, the extent to which disease-related variables such as disease severity and type of treatment affect perceived competence has not yet been

addressed, nor has the role of demographic variables or joint status been investigated in relation to these variables.

Another psychological aspect of importance may include the so-called perceived impact of disease [17,18]. Children with haemophilia may differ in their understanding of the impact of the disease and its potential limitations. Until now, it remains unknown to what extent children with haemophilia feel that their lives are compromised by the social implications of their illness. In addition, it can be hypothesized that patients who experience a high negative impact of their disease also perceive lower competence.

The aims of the present study were to explore: (i) the level of perceived competence and (ii) the perceived impact of disease in children and adolescents with haemophilia. A third aim was to analyse associations between perceived competence and demographic factors (age, education), disease-related factors (type, severity and treatment), joint status (pain and range of motion) and perceived impact of disease in children and adolescents with haemophilia in the Netherlands. A final goal of the study was to obtain possible tools or suggestions for intervention. Therefore, we aimed to provide insight into the association between perceived competence and functional status in addition to the variables mentioned above.

## Methods

### *Procedure*

This study is part of a larger trial [19,20] that included a cross-sectional, multi-centre, explorative investigation. The study was performed in five haemophilia treatment centres in the Netherlands (Amsterdam, Groningen, Nijmegen, Rotterdam and Utrecht). Patients (and their parents) under care and supervision of the multidisciplinary haemophilia team of these five centres were informed about the study by phone and an information letter. Subsequently, informed consent was obtained from parents and/or the participating child. The study was approved by all institutions' medical ethics committees.

The study comprised a physical assessment in the haemophilia centres and an assessment by questionnaires. Physical examination of the population of the treatment centre in Utrecht was performed in 2006 and the questionnaires were gathered in 2007. The children in the other four centres completed questionnaires and the physical examination between March and October 2007.

Inter-rater agreement between the paediatric physiotherapists of the participating centres regarding joint mobility was determined in four healthy boys assessed by five raters prior to the study and appeared to be acceptable (Intraclass Correlation Coefficient  $\geq 0.80$ ).

### Participants

All boys with haemophilia A or B (regardless of disease severity), aged 8–18 years, treated at one of the five participating centres were considered for the present study. Patients were excluded from the study if they (i) had insufficient understanding of Dutch language (spoken and written), (ii) suffered from acute pathology (muscle or joint bleeding) during the 2 weeks prior to the physical assessment, or (iii) had relevant co-morbidities such as Down syndrome, psychomotor retardation or epilepsy.

In total, 243 patients were eligible for the study, 158 (65%) of whom participated in the main study and the physical assessments. Analysis of the responders vs. the non-responders revealed that the responders were significantly younger than the non-responders (12.7 vs. 13.9 years ( $P = 0.003$ ), respectively). No significant differences were found regarding diagnosis (i.e. haemophilia type and severity) or z-scores for height, weight and body mass index ( $P \geq 0.05$ ).

Completed questionnaires on the main outcomes of the present sub-study were available for 126 patients (80% of the responders), including 54 children (age 8–12 years) and 72 adolescents (12–18 years). Therefore, the completion rate for the present study was 52% (126/243) and these data were considered for further analysis. No significant differences between patients who did ( $n = 126$ ) and did not ( $n = 32$ ) complete the questionnaires were found with respect to age, haemophilia type and severity, treatment modality and level of education ( $P \geq 0.05$ ). Outcomes of the functional status [i.e. 6-min walk test (6MWT)] were available in a subgroup of 41 children and 44 adolescents.

### Measures

1. *Demographic variables* including age and level of education were administered.
2. *Disease-related variables* including type (haemophilia A or B) and severity [mild (5–40% clotting activity), moderate (1–5%) and severe (<1%)] and medical treatment (on demand or prophylaxis) were taken from the medical chart.
3. *Joint status [i.e. pain and active range of motion (AROM)]* was examined by physical assessment
  - a. *Pain* was assessed in six joints (i.e. elbows, knees and ankles bilaterally). Patients were asked whether or not they had painful joints. Subsequently, for each painful joint, a Visual Analogue Scale was administered to quantify to what extent they suffered from painful joints. The total number of painful joints was taken for analyses.
  - b. *Joint mobility*. The AROM of the elbow (flexion and extension), knee (flexion and extension) and ankle joint (plantar- and dorsal extension) was measured bilaterally using a standard 2-legged 360°

goniometer (Sulzer Medica, Bern, Switzerland), because these joints are the most frequently affected in haemophilia. Children were asked to actively stretch or bend the joint maximally without interference by the investigator. For each child, a Total AROM was calculated by summing up the AROM from all measured joints; this score was used for the analyses and compared with normal values (i.e. 710°) [21]. In addition, the overall AROM was recalculated into a z-score based on reference scores of a normative population [22], consisting of 118 boys, with a mean age of 12.9 years (SD 3.3).

4. *Perceived competence* was measured with the Dutch version [10,23] of the Self Perception Profile for Children (SPPC) [24]/Adolescents (SPPA) [25] questionnaire. The SPPC contains 36 items for five domain-specific subscales of perceived competence (i.e. scholastic competence, social acceptance, athletic competence, physical appearance and behavioural conduct) and one global self-worth subscale. The SPPA contains 40 items for six domain-specific subscales (with an additional domain for close friendship) and one global self-worth scale. In the questionnaire, respondents were asked to choose between two opposing statements and indicate whether that statement is 'somewhat true' or 'very true' for their specific situation. The total score per subscale for children ranges from 6 to 24 and for adolescents from 5 to 20; higher scores refer to a higher perceived competence. Cronbach's alpha for children and adolescents on the different subscales in the present study ranged from 0.52 to 0.77. The Dutch version of the SPPC and SPPA provide reference scores of Dutch children ( $n = 180$  boys, age 8–12 years) [23] and adolescents ( $n = 601$  boys, age 12–18 years) [10].
5. Besides perceived competence, the questionnaire provides two domain *importance* questions for the cognitive, social, motor, appearance and behavioural conduct domains. These domain importance scores range from 1 to 4, and a domain with a score  $\geq 3$  is classified as being important to the child. Furthermore, *discrepancy scores* (i.e. the difference between importance and judged competence per domain) can be calculated to reflect the child's sense of self-worth. The higher the discrepancy in negative direction (i.e. the more one's importance scores exceeds on one's competency scores), the lower one's self-worth is. The closer the discrepancy score moves to zero the higher the self-worth [8,9].
6. *Impact of disease* was measured using a Dutch translation [26,27] of the Revised Perception Illness Experience (R-PIE) questionnaire [17,18]. The R-PIE consists of 24 items that include a statement on the impact of the disease on daily life regarding the following six subscales: school/peer rejection, thinking about illness, physical appearance,

interference with activity, parental responses and manipulation. Examples of statements are 'I find it hard to learn things because of my disease', and 'My disease is an impediment to partake in games or sports that I like'. Children rate the extent to which they agree or disagree with the statement on a 5-point Likert scale (1 = fully disagree to 5 = fully agree). A composite R-PIE score was calculated by summing all six subscale scores and dividing this by six. Higher scores indicate more negative illness experiences and thus, a higher negative impact. Cronbach's alpha for the present study ranged from 0.39 to 0.72 for the subscales, and an  $\alpha = 0.80$  for the composite R-PIE.

7. *Functional status* was assessed in a subpopulation (i.e. 41 children and 44 adolescents) of the larger trial by the 6MWT. The 6MWT measures the distance that a child can walk without running on a flat and hard surface in 6 min. The children were encouraged to walk as far as possible in 6 min. The walking course was of 20–30 m in length [28]. The children were kept informed about the progress of time. The maximum walked distance in metres and a z-score walking distance for age were used for analysis [29].

### Statistics

Statistical analysis was performed using SPSS, version 14.0 (SPSS Inc., Chicago, IL, USA); *t*-tests and chi-squared tests were used to assess differences in socio-demographic and medical characteristics between responders and non-responders in this study. Mann-Whitney *U*-tests were performed to assess differences between patients with mild, moderate and severe disease. Scores of AROM and 6MWT were transformed to standard deviation scores (z-scores), i.e. measured outcome – mean outcome in the reference group/SD in the reference group. For the AROM outcomes, reference values were only available for the whole group [20], whereas reference values for the 6MWT were available per age group [29]. One-sample *t*-tests (with test value = 0) were applied to assess significant differences between the study population and the reference groups, and frequencies were used to explore the percentage of the study population that exceeded the limits of +2 and –2. Confidence Interval Analyses [30] were performed to compare perceived competence of the patients with reference groups of the Dutch population [10,23]. For all tests,  $\alpha < 0.05$  (two-tailed) was considered statistically significant. Correlation analyses (and *t*-tests) were performed to explore the association (differences) between perceived competence on one hand and demographic, disease-related factors, joint status and perceived impact of disease on the other. In addition, correlation analyses between perceived competence and functional status were performed for the subpopulation. Bonferroni corrections [31] were

applied to correct for multiple testing and to prevent type I errors. Therefore,  $\alpha$  was divided by the number of variables used in the model, i.e.  $0.05/6 = 0.008$ . Kruskal–Wallis tests were used to assess differences between the centres.

## Results

### Patient characteristics

Mean (SD) age of children (8–12 years) and adolescents (12–18 years) was 10.0 (1.0) and 15.0 (1.6) years, respectively. Most children (89%) and adolescents (88%) had haemophilia type A. In children, 46% had mild, 11% moderate and 43% severe haemophilia. In adolescents, 43% had mild, 17% moderate and 40% severe haemophilia. In children, 52% had medication on demand and 48% were on prophylaxis. In adolescents, 63% and 37% had treatment on demand and prophylactic medication, respectively (Table 1). Chi-squared tests showed a significant association between disease severity and medication; 92% of the children and 100% of the adolescents with a mild disease received on-demand treatment, whereas all children and 86% of the adolescents with severe disease were on prophylaxis ( $\chi^2 = 43.3$ ,  $P < 0.001$  and  $\chi^2 = 50.2$ ,  $P < 0.001$ , respectively). Most children and adolescents (83% in both groups) reported to have no painful joints. Seven percent of the children reported to have one

**Table 1.** Patients' characteristics for children and adolescents.

	Children 8 to ≤12 years <i>n</i> = 54	Adolescents >12–18 years <i>n</i> = 72
Mean (SD) age in years	10.0 (1.0)	15.0 (1.6)
Level of education ( <i>n</i> , %)		
Primary	52 (96.3)	2 (2.8)
Lower and middle vocational education	–	33 (45.8)
Secondary education	2 (93.7)	35 (48.6)
Higher vocational/university	–	1 (1.4)
Missing	–	1 (1.4)
Type of haemophilia ( <i>n</i> , %)		
A	48 (88.9)	63 (87.5)
B	6 (11.1)	9 (12.5)
Severity ( <i>n</i> , %)		
Mild	25 (46.3)	31 (43.0)
Moderate	6 (11.1)	12 (16.7)
Severe	23 (42.6)	29 (40.3)
Treatment		
On demand	28 (51.9)	45 (62.5)
Prophylaxis	26 (48.1)	27 (37.5)
Painful joints ( <i>n</i> , %)		
No	45 (83.3)	60 (83.3)
Yes	9 (16.7)	12 (16.7)
Mean (SD) AROM	740.1 (36.6)	717.5 (41.7)
Mean (SD) z-score	0.27 (0.53)	–0.03 (0.59)
Mean (SD) walking distance (Mtr)	680 (100.2)	691 (82.7)
Mean (SD) z-score for age	0.39 (1.4)	–0.2 (1.2)

SD, standard deviation; AROM, active range of motion, Mtr, metre.



( $n = 4$ ), two ( $n = 3$ ), three ( $n = 1$ ) and four ( $n = 1$ ) painful joints, and adolescents reported to have one ( $n = 8$ ) and two ( $n = 4$ ) painful joints. Mean (SD) range of motion for children and adolescents was 740 (37)° and 718 (42)° respectively. Mean (SD) z-scores for AROM were 0.27 (1.40) for children, and -0.03 (0.59) for adolescents. One-sample *t*-tests of z-scores showed a significantly higher joint mobility in the children with haemophilia compared with the reference group ( $P < 0.001$ , CI 0.12 to 0.42). No significant differences were found between adolescents with haemophilia and a reference group ( $P > 0.05$ , CI -0.17 to 0.11).

In subgroup analyses (i.e. 41 children and 44 adolescents), the mean (SD) for walking distance metres in children and adolescents were 680 (100) and 691 (83) m, respectively. Mean (SD) z-scores for walking distance for age were 0.39 (1.5), and -0.02 (1.2), respectively. One-sample *t*-tests of z-scores revealed no significant differences between the children and adolescents with haemophilia and a normal population [29] ( $P > 0.05$ ). *Post hoc* Kruskal-Wallis analyses showed significant differences between the centres.

### Perceived competence

In children, mean (SD) scores for perceived competence ranged from 17.0 (4.0) to 19.6 (4.0) and for adolescents from 13.3 (2.4) to 15.7 (2.8). No significant differences were found between the children with haemophilia and the Dutch reference group of healthy children [10] except a lower global self-worth [18.9 (4.0) vs. 20.0 (3.0) (mean (SD),  $P \leq 0.01$ ; CI 0.11–2.09)]. Adolescents with haemophilia had a significantly lower perceived competence for close friendship when compared with the Dutch reference group [23] [15.6 (2.8) vs. 16.6 (2.9) (mean (SD);  $P \leq 0.01$ , CI: 0.29–1.71)] but no significant differences were found for the other domains (Table 2). Importance scores revealed that 94%, 63%, 69%, 52% and 58% of the children rated the cognitive, social, motor, physical appearance and behavioural conduct

domains as being important respectively. For adolescents, the corresponding percentages were 90%, 60%, 68%, 60% and 89% respectively. In children and adolescents, the discrepancy scores (i.e. the difference between importance and judged competence per domain) ranged from -0.04 to -0.60 and from -0.07 to -0.72, respectively, indicating normal levels of self-worth in both groups. *Post hoc* Kruskal-Wallis analyses showed significant differences in perceived competence between the centres.

### Impact of disease

Mean (SD) scores for perceptions of impact of disease ranged from 1.2 (0.4) to 2.2 (0.9) in children and from 1.3 (0.4) to 2.0 (0.8) in adolescents, with the highest scores (i.e. most negative impact) on thinking about the disease and interference with activities. Scores on the composite R-PIE were 1.8 (0.4) and 1.7 (0.5) respectively, all indicating a low negative impact of the disease (Table 3).

### Associations between perceived competence and socio-demographic, disease-related factors, joint status and perceived impact of disease in children and adolescents

**Children.** No associations were found between age and perceived competence. Significant associations were found between disease severity and five domains of perceived competence (Table 4). Children with severe disease (correlation analyses) or prophylactic treatment (Mann-Whitney *U*-tests) had significantly lower scores on school competence, athletic competence, physical appearance, behavioural conduct and global self-worth than children with mild disease. Besides the finding that a larger AROM was associated with a lower physical appearance, no associations were found between joint status (pain and range of motion) and perceived competence. In addition, a high perceived impact of

**Table 2.** Scores of perceived competence for children (SPPC) and adolescents (SPPA) with haemophilia and general populations and Confidence Interval Analyses (CIA).

	Children Mean (SD)	General population Mean (SD)	Adolescents Mean (SD)	General population Mean (SD)
Scholastic competence	17.3 (4.0) Range (7–24)	17.4 (3.5)	14.0 (3.0) Range (7–19)	14.5 (2.5)
Social acceptance	18.2 (4.2) Range (11–24)	17.8 (3.8)	14.9 (2.7) Range (9–20)	15.3 (2.7)
Athletic competence	17.7 (3.5) Range (11–24)	18.7 (3.3)	14.1 (2.9) Range (8–20)	14.8 (3.3)
Physical appearance	19.6 (4.0) Range (12–24)	20.1 (3.6)	14.7 (3.0) Range (7–20)	14.7 (3.1)
Behavioural conduct	17.3 (3.6) Range (10–24)	17.0 (2.8)	13.3 (2.4) Range (8–19)	13.8 (2.8)
Close friendship	–	–	15.6 (2.8)* Range (7–20)	16.6 (2.9)
Global self-worth	18.9 (4.0)* Range (11–24)	20.0 (3.0)	15.7 (2.8) Range (9–20)	16.0 (2.7)

SD, standard deviation; SPPC, Self Perception Profile for Children; SPPA, Self Perception Profile for Adolescents. \* $P < 0.05$ , comparisons based on confidence interval analysis.

**Table 3.** Scores of perceived impact of disease (R-PIE) for children and adolescents with haemophilia.

	Children Mean (SD)	Adolescents Mean (SD)
Peer rejection	1.7 (0.6)	1.5 (0.5)
Thinking	2.3 (0.8)	2.0 (0.8)
Physical appearance	1.2 (0.4)	1.3 (0.4)
Interference activities	2.2 (0.9)	2.0 (0.8)
Parental responses	1.6 (0.6)	1.8 (0.7)
Manipulation	1.5 (0.6)	1.7 (0.8)
Composite R-PIE	1.8 (0.4)	1.7 (0.5)
	Range (1.1–2.9)	Range (1.1–3.1)

SD, standard deviation; R-PIE; Revised Perception Illness Experience scale.

disease was associated with lower scores on social acceptance, physical appearance, behavioural conduct and global self-worth. In a subgroup of 41 children, supplemental correlation analyses between functional status and perceived competence revealed that a shorter walking distance was associated with lower levels of school competence, athletic competence, physical appearance, behavioural conduct and global self-worth.

**Adolescents.** Apart from the finding that older patients showed higher scores on school competence ( $r = 0.25$ ,  $P = 0.04$ ), no further associations were found between age and perceived competence. ANOVA revealed that adolescents with secondary education showed lower scores on physical appearance and global self-worth compared with adolescents with lower or middle vocational education, and a trend was also found for social acceptance ( $P = 0.07$ ) and close friends ( $P = 0.09$ ). In contrast to children, no significant associations were found between disease severity and perceived competence neither in adolescents nor between treatment modality and perceived competence. In accordance with the findings in children, no associations were found between joint status, i.e. pain and range of motion, and perceived competence. Finally, a high impact of disease was significantly associated with lower scores on global self-worth only. In a subgroup of 44 adolescents, correlation analyses between functional status and perceived competence revealed that a shorter walking distance was significantly associated with lower scores on social acceptance, athletic competence and close friends, while a trend was found for global self-worth ( $P = 0.09$ ).

## Discussion

The results of this multi-centre study in the Netherlands show that children and adolescents with haemophilia in general have a perceived competence that is comparable with that of healthy peers, with the exception of a lower

**Table 4.** Univariate non-parametric correlations (Spearman's rho) and Mann-Whitney *U*-tests between perceived competence and disease-related factors (and functional status) in children and adolescents.

	Scholastic competence		Social acceptance		Athletic competence		Physical appearance		Behavioural conduct		Global self-worth		Close friends	
	Children	Adolescent	Children	Adolescent	Children	Adolescent	Children	Adolescent	Children	Adolescent	Children	Adolescent	Children	Adolescent
Disease severity	<b>-.46**</b>	.08	-.17	-.12	<b>-.38**</b>	-.13	-.31*	.07	<b>-.42**</b>	.01	<b>-.38**</b>	.10	-.05	
Treatment	<b>-2.99***</b>	-.54	-.65	-.67	<b>-2.15*</b>	-.11	<b>-2.27*</b>	-.94	<b>-2.43*</b>	-.65	<b>-2.01*</b>	-.33	-.20	
Pain	-.03	.17	.19	-.09	.06	.11	.14	-.03	.14	.16	.15	-.00	-.03	
Active range of motion	-.12	-.02	-.05	-.11	-.13	.10	-.30*	.05	-.04	-.03	-.20	.04	-.04	
Composite impact of disease	-.09	-.14	-.31*	-.06	-.19	-.00	<b>-.43**</b>	.10	-.33*	.02	<b>-.29*</b>	-.08	.04	
Functional status	.34*	.25	.27	.36*	<b>.61***</b>	<b>.41**</b>	<b>.65***</b>	.13	.34*	.14	<b>.57***</b>	.26	.39*	

\*  $P < 0.05$ , \*\*  $P < 0.01$ , \*\*\*  $P < 0.001$ . Bold: significant after Bonferroni corrections.

global self-worth in children and a lower perceived competence in close friendship for adolescents. Furthermore, children and adolescents with haemophilia perceive their disease as having relatively low impact on their life. Univariate explorations – with Bonferroni corrections – showed that severe disease, prophylactic medication and high perceived impact of disease were associated with lower perceived competence in children. Additional explorations in subgroups revealed that a lower functional status was associated with lower perceived competence.

An important finding regarding the first aim of the study was that, in our sample, children with haemophilia showed levels of perceived competence that were nearly comparable with the normative population [10]; they appear to perceive themselves as competent as their unaffected peers across a range of skill areas. Moreover, the low discrepancy scores show that competence and importance hierarchies are quite congruent, indicating a high self-worth in five domains. Thus, despite the presence of a chronic disease, children with haemophilia show a normal perceived competence. However, the study revealed a significantly lower global self-worth in children with haemophilia, which may require further attention. The global self-worth scale constitutes a global judgment of one's worth as a person rather than domain-specific competence or adequacy [8,32]. It represents 'the ratio of one's successes to one's pretensions towards success in the various domains of one's life', as well as 'the reflected appraisals of significant others'. The first part may indicate that children with haemophilia have less successful experiences compared with those of their healthy peers, whereas the second part may indicate that they have an environment that is not stimulating or responsive enough because of a lack of social and/or emotional support [8]. In both cases, an overprotective attitude of parents may aggravate a lower global self-worth. This is underlined by other studies reporting associations between high parenting stress and low self-concept among children with epilepsy [13]. Therefore, it might be useful to provide parents with adequate education about haemophilia and to emphasize the need of social emotional support including motivating children to perform activities and thereby being able to perceive success.

Similar to the children in our study, adolescents with haemophilia reported mean levels of competence that were relatively consistent with those of the normative standardized group [23], with the exception of a lower score on close friendship. The relatively high ratings of self-competence may not be surprising in light of the low number of complaints reported by patients. However, adolescents did feel less confident in making friends compared with the normative sample. When combining this finding with the relatively high importance rating given to social acceptance, one has to address the importance of external factors including the adolescents'

environment. As the typical developmental tasks of adolescence include forming a cohesive self-identity, enhancing closeness and trust with peers, and redefining relationships with parents [33], it could be worthwhile to discuss these subjects in adolescents with haemophilia.

The results found in this study were partially in accordance with a previous pilot study involving children in the age of 8–15 years in the United States [15]. That study found significant lower scores on global self-worth, and on the domains of scholastic and athletic competence in boys with haemophilia. Differences with our study may be due to cultural differences between the Dutch and the American population or selection bias as the non-responders in our study were significantly older than the responders. The lower score on global self-worth found in both studies underlines, as previously discussed, the need for attention and information. The merit of our study is that we included a large sample size and used age-specific instruments to measure perceived competence in children and adolescents separately.

The second aim of our study was to explore the perceived impact of disease in young patients with haemophilia. Regarding the R-PIE, the interpretation of the results should be taken with caution because the Cronbach's alpha for some of the subscales were insufficient, indicating a low degree of internal consistency. The results of the R-PIE suggest that both children and adolescents with haemophilia do not perceive a strong negative impact of their disease on their lives. We found that the impact of disease was the highest on thinking and interference with activities. The higher scores on thinking might indicate that patients with haemophilia wish to be treated as healthy persons without being reminded of their illness by their social environment. The score on perceived interference with activities may be associated with the need of medication, pain or potential limitations in sport participation. However, pain was not an issue in the study population. Moreover, physical limitations were not present as well, as AROM and functional status were higher or within normal ranges. Therefore, it may be that perceived interference with games and activities are based on needed medication or imposed by restraints through the environment (i.e. parents, teachers), or due to a difference between patients' perception and practising of sports.

Regarding the third aim, our study revealed that perceived competence in children with haemophilia was associated with disease-related variables. Children with severe haemophilia had significantly lower scores on perceived competence than patients with mild disease. Furthermore, prophylactic medication was associated with lower perceived competence, although this can probably be attributed to a strong association between disease severity and treatment. After all, in case of severe haemophilia, patients do have a medical treatment consisting of injecting two or three times a week, which may negatively affect competency judgements.



The study further showed that the joint status was not associated with perceived competence, but a negative association was found between perceived impact of the disease and perceived competence. Thus, while objective parameters did not affect perceived competence, it seemed that those who experienced a higher negative impact of the disease perceived lower levels of competence. This might be in accordance with the notion that varied experiences of failure and success generate feelings of competence [34,35].

This study finally showed in subgroup analyses that functional status was associated with perceived competence, i.e. lower walking distance was associated with lower scores on athletic competence, physical appearance and global self-worth in children, and with lower scores on social acceptance and athletic competence in adolescents. This indicates that those children and adolescents with lower physical functioning may be at risk for lower perceived competence in several domains or vice versa. One may hypothesize that children with haemophilia may (i) have less *successful* experiences or (ii) have a *lack of experiences* in the physical domain. Moreover, a vicious circle of decreased competence and low performance may occur. To break through this circle, the environment should be informed and stimulated to give their children the opportunity to perceive (mastery) experiences in physical activities.

#### *Limitations and strengths of the study*

A limitation of the study may be the possibility of selection bias. Results can be biased because, since perceived competence is associated with age, the responders were significantly younger than the non-responders. Furthermore, the results concerning the walking distance should be considered with caution because centre differences were found in our study. It appeared that, although the 6MWT is a standardized test, the walking course in one of the centres – exhibiting the highest scores on walking distance – was not identical to the others, and children in that centre inadvertently received extra encouragement. Moreover, reference values for the 6MWT – as no reference values for Dutch children are available – were obtained from an Austrian population that walked with a measuring wheel [29]. Although the interpretation of the comparison of the walking distance between our population and the normal population should be read with caution, these restrictions do not affect the found association between walking distance and perceived competence. A final limitation of the study might be the generalization beyond countries in Western Europe, as our study was conducted in the Dutch population.

The strength of our study was that 65% of all children and adolescents with haemophilia in the Netherlands participated in the physical assessments of the study.

Eighty percent of them completed questionnaires on perceived competence. Thus, we included a large and representative sample of the Dutch population. Although data regarding the functional status were only obtained in a subgroup (i.e. two-third of the population), this is the most comprehensive study on perceived competence in young haemophiliacs in the Netherlands so far. An additional merit of our study was that we distinguished between children and adolescents.

#### *Practical implications*

Patients with severe haemophilia and/or low functional status may be at risk of lower perceived competence or vice versa. Therefore, screening on perceived competence might be incorporated in the usual care for haemophilia and one can recommend some strategies to enhance perceived competence for those with lower scores. Such strategies may include adequate information for patients and their environment or interventions that make use of mastery experiences, vicarious experiences, verbal persuasion and reinterpretation of physiological and affective state [34]. Such interventions may be psycho-educational in nature [36,37], but may also include physical interventions. Physical interventions have the merit that the successful accomplishment of physical exercise itself (i.e. mastery experience) may result in a positive effect on patients' perceived competence [38].

#### **Conclusion**

Children and adolescents with haemophilia in general have a perceived competence that is comparable with that of healthy peers, with the exception of a lower global self-worth in children and a lower competence in close friendship for adolescents. Furthermore, they seem to perceive their disease as having relatively low impact on their life. Severe disease, prophylactic treatment and low functional status seem to be associated with lower perceived competence.

#### **Acknowledgements**

We would like to thank the patients and their parents for participating in this study. The Hemofit Collaborative Study Group consists of M. Peters MD PhD, M. Beijleveld - Van der Zande, A.C.M. Beishuizen - Van Remmen (AMC, Amsterdam); D. Douma - Van Riet, A. Hartman MSc (Erasmus MC, Rotterdam); J. Koiter, M.W.G. Nijhuis - Van der Sanden PhD (UMC St Radboud, Nijmegen); and R.H.H. Engelbert PhD, J. Van der Net PhD (UMC Utrecht, Utrecht); and was supported by an unrestricted grant from Baxter B.V., the Netherlands.

#### **Disclosures**

The study was supported by an unrestricted grant from Baxter B.V., Utrecht, The Netherlands. The authors stated that they had no interests which might be perceived as posing a conflict or bias.

## References

- Lofqvist T, Nilsson IM, Berntorp E, Pettersson H. Haemophilia prophylaxis in young patients—a long-term follow-up. *J Intern Med* 1997; **241**: 395–400.
- Van Der Net J, Vos RC, Engelbert RH, Van den Berg MH, Helden PJ, Takken T. Physical fitness, functional ability and quality of life in children with severe haemophilia: a pilot study. *Haemophilia* 2006; **12**: 494–9.
- Engelbert RH, Plantinga M, Van Der Net J *et al*. Aerobic capacity in children with haemophilia. *J Pediatr* 2008; **152**: 833–8.
- Falk B, Portal S, Tiktinsky R, Weinstein Y, Constantini N, Martinowitz U. Anaerobic power and muscle strength in young haemophilia patients. *Med Sci Sports Exerc* 2000; **32**: 52–7.
- Falk B, Portal S, Tiktinsky R *et al*. Bone properties and muscle strength of young haemophilia patients. *Haemophilia* 2005; **11**: 380–6.
- Koch B, Galioto FM Jr, Kelleher J, Goldstein D. Physical fitness in children with haemophilia. *Arch Phys Med Rehabil* 1984; **65**: 324–6.
- Schoenmakers MA, Gulmans VA, Helden PJ, Van Den Berg HM. Motor performance and disability in Dutch children with haemophilia: a comparison with their healthy peers. *Haemophilia* 2001; **7**: 293–8.
- Harter S. Causes, correlates, and the functional role of global self-worth. A life-span perspective. In: Sternberg RJ, Kolligian J eds. *Perceptions of competence and in competence across the life-span*. New Haven: Yale University Press, 1990: 136–82.
- Harter S. Processes underlying the construction, maintenance and enhancement of self concept in children. In: Suls J, Greenwald AG eds. *Psychological Perspectives on the Self*. Hillsdale: Lawrence Erlbaum, 1986: 136–82.
- Veerman JW. *Competentie Belevingsschaal voor Kinderen*. Lisse: Swets & Zeitlinger, 1997.
- Miatton M, De Wolf D, Francois K, Thiery E, Vingerhoets G. Behavior and self-perception in children with a surgically corrected congenital heart disease. *J Dev Behav Pediatr* 2007; **28**: 294–301.
- Guite JW, Logan DE, Sherry DD, Rose JB. Adolescent self-perception: associations with chronic musculoskeletal pain and functional disability. *J Pain* 2007; **8**: 379–86.
- Chiou HH, Hsieh LP. Comparative study of children's self-concepts and parenting stress between families of children with epilepsy and asthma. *J Nurs Res* 2008; **16**: 65–74.
- Russo RN, Miller MD, Haan E, Cameron ID, Crotty M. Pain characteristics and their association with quality of life and self-concept in children with hemiplegic cerebral palsy identified from a population register. *Clin J Pain* 2008; **24**: 335–42.
- Trzepacz AM, Vannatta K, Davies WH, Stehbens JA, Noll RB. Social, emotional, and behavioral functioning of children with haemophilia. *J Dev Behav Pediatr* 2003; **24**: 225–32.
- Veerman JW, Ten Brink LT, Straathof MA, Treffers PD. Measuring children's self-concept with a Dutch version of the "self-perception profile for children": factorial validity and invariance across a nonclinic and a clinic group. *J Pers Assess* 1996; **67**: 142–54.
- Eiser C, Kopel S, Cool P, Grimer R. The Perceived Illness Experience Scale (PIE): reliability and validity revisited. *Child Care Health Dev* 1999; **25**: 179–90.
- Kiernan G. The Revised-Perceived Illness Experience Scale (R-PIE): data from Barretstown studies. *Behav Med* 2004; **30**: 23–31.
- Douma-Van Riet DC, Engelbert RH, Van Genderen FR, Ter Horst-De Ronde MT, Goede-Bolder A, Hartman A. Physical fitness in children with haemophilia and the effect of overweight. *Haemophilia* 2009; **15**: 519–27.
- Koiter J, Van Genderen FR, Brons PP, Nijhuis-van der Sanden MW. Participation and risk-taking behaviour in sports in children with haemophilia. *Haemophilia* 2009; **15**: 686–94.
- Soucie JM, Cianfrini C, Janco RL *et al*. Joint range-of-motion limitations among young males with haemophilia: prevalence and risk factors. *Blood* 2004; **103**: 2467–73.
- Engelbert RH, Uiterwaal CS, van de Putte E *et al*. Pediatric generalized joint hypomobility and musculoskeletal complaints: a new entity? Clinical, biochemical, and osseal characteristics. *Pediatrics* 2004; **113**: 714–9.
- Treffers PhDA. *Competentie Belevingsschaal voor Adolescenten (CBSA)*. Lisse: Swets & Zeitlinger, 2000.
- Harter S. *Manual for the Self-Perception Profile for Children*. Denver: University of Denver, 1985.
- Harter S. *Manual for the Self-Perception Profile for Adolescents*. Denver: University of Denver, 1988.
- Brislin R. Back-translation for cross-cultural research. *Journal of Cross Cultural Psychology* 1970; **3**: 185–216.
- Brislin R. *The Wording and Translation of Research Instruments. Field Methods in Cross-Cultural Research. Cross-Cultural Research and Methodology Series*. Thousand Oaks: Sage Publications, 1986.
- ATS Board of Directors. ATS statement: guidelines for the six-minute walk test. *Am J Respir Crit Care Med* 2002; **166**: 111–7.
- Geiger R, Strasak A, Trembl B *et al*. Six-minute walk test in children and adolescents. *J Pediatr* 2007; **150**: 395–9.
- Altman D, Machin D, Bryant T, Gardner S. *Statistics with Confidence*. 2nd edn. Oxford, UK: Blackwell BMJ Books, 2008.
- Field A. *Discovering Statistics Using SPSS*. London: SAGE Publications Ltd, 2005.
- Harter S. Developmental Differences in the Nature of Self-Representations: Implications for understanding, assessment, and Treatment of Maladaptive Behavior. *Cognit Ther and Res* 1990; **14**: 113–42.
- Sroufe LA, Cooper RG. Social development in adolescence. In: Sroufe LA, Cooper RG, Marshall ME eds. *Child Development: Its Nature and Course*. New York: Knopf, 1998: 608.
- Bandura A. Self-efficacy. In: Ramachandran VS ed. *Encyclopaedia of Human Behaviour*. New York: Academic Press, 1994: 71–81.
- Bandura A. Self-Efficacy - Toward A Unifying Theory of Behavioral Change. *Psychol Rev* 1977; **84**: 191–215.
- Lorig K, Lubeck D, Kraines RG, Seleznick M, Holman HR. Outcomes of self-help education for patients with arthritis. *Arthritis Rheum* 1985; **28**: 680–5.
- Taal E, Riemsma RP, Brus HL, Seydel ER, Rasker JJ, Wiegman O. Group education for patients with rheumatoid arthritis. *Patient Educ Couns* 1993; **3**: 177–87.
- Gulmans VA, De Meer K, Brackel HJ, Faber JA, Berger R, Helden PJ. Outpatient exercise training in children with cystic fibrosis: physiological effects, perceived competence, and acceptability. *Pediatr Pulmonol* 1999; **28**: 39–46.